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Sir,

Surgically induced necrotising scleritis: report of a case presenting 51 years following strabismus surgery

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Necrotising scleritis is a destructive inflammatory disease of the sclera with serious ocular complications including keratitis, peripheral corneal ulceration and perforation with poor visual prognosis. It is usually associated with systemic diseases in particular the collagen vascular disorders and autoimmune conditions. Surgically induced scleral necrosis (SINS) is a rare form of necrotising scleritis occurring at a site of previous ocular surgery.

We report a case of unilateral necrotising scleritis and peripheral ulcerative keratitis occurring 51 years after ocular surgery. Forty years is the previously longest reported interval.[1] Systemic immunosuppression was required to stabilize the progressive necrosis. Healing was promoted with the use of a silicone hydrogel bandage contact lens.

Case report

A 59-year-old Caucasian woman presented with a history of episodic pain in her left eye and a small dark spot developing on the nasal side of the sclera over a period of 12 months. Her past medical history consisted only of two strabismus operations at the age of 7 and 8 on the affected eye. No written records were available. At initial presentation her visual acuity was 6/6 in both eyes. A focal area of scleral thinning with localised injection on the nasal side of the left eye was noted. The right eye was normal.

Following the diagnosis of anterior scleritis the patient was commenced on topical ketorolac and prednisolone acetate 1% which resulted in moderate symptomatic improvement after 2 months. Oral flurbiprofen was added to help with symptoms but on further review 2 months later, the scleral lesion was increasing in size with recurrent and persistent discomfort in the eye. At this stage there was increased thinning of the sclera with the underlying uveal tissue appearing more prominent. The adjacent peripheral cornea showed an area of guttering associated with loss of epithelium (Figure 1). Careful examination of the bulbar conjunctiva over the lateral rectus revealed subconjunctival scarring consistent with previous surgery. There was no evidence of retained suture material.

A revised diagnosis of necrotising anterior scleritis was made at this stage. Systemic immunosuppression was commenced in the form of a pulse of methylprednisolone 500 mg followed by prednisolone 20 mg, reduced to 10 mg maintenance dose within a week. Topical prednisolone phosphate was continued.

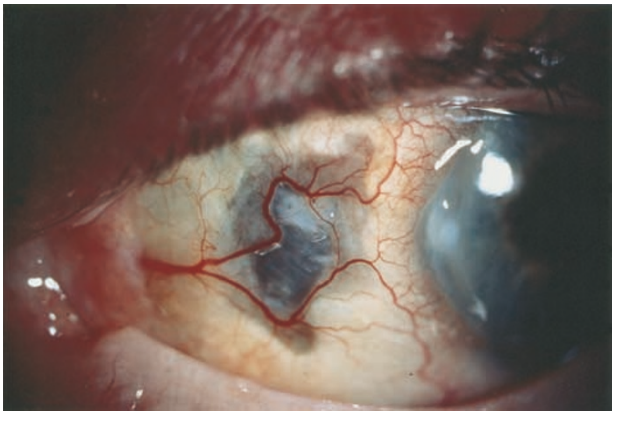

Figure 1 Medial view of the left eye showing a large area of scleral thinning at the site of previous ocular surgery with associated peripheral ulcerative keratitis.

A general physical examination was performed before immunosuppression by a rheumatologist (INB) but there was no evidence of systemic vasculitis or arthritis. A chest x-ray and blood tests including a full blood count, ESR, CRP, rheumatoid factor, ANA and ANCA were performed which were found to be normal. Urinalysis was normal.

There was initial symptomatic improvement and the size of the lesion did not increase further on systemic steroid. Prednisolone was continued at 10 mg because no significant healing was noted. However over a period of 5 months there was a gradual recurrence of pain. Two further pulses of methylprednisolone were given without success. The patient was then fitted with a silicone hydrogel bandage contact lens which was highly effective in relieving symptoms, and at 2 months the lesion had significantly decreased in size and corneal guttering healed. After a further 3 months her prednisolone had been tailed down to 2.5 mg and at the time of writing she remains comfortable wearing the contact lens with continued slow healing of her lesion.

Comment

Surgically induced necrotising scleritis (SINS) may be defined as scleral inflammation with necrosis occurring after ocular surgery. It most commonly occurs at the incision site for cataract surgery and following strabismus surgery. In the largest review of 52 eyes by O'Donoghue *et al*,[1] 75% of patients had two or more ocular procedures including bilateral cataract extraction, cataract extraction followed by secondary ocular surgery and multiple detachment procedures. The scleral necrosis rarely occurs in the immediate postoperative period. The mean interval was 9 months after multiple procedures compared with 9.5 years after a single ocular procedure. The previously longest reported interval was 40 years.[1]

An autoimmune mechanism has been postulated, either a delayed type hypersensitivity reaction against an antigen revealed or altered by tissue injury, or the process may represent molecular mimicry or cross reactivity between an ocular antigen and another body tissue or microbial antigen. Nine out of ten patients with SINS reported by De la Maza *et al*[2] and 63% of patients reviewed by O'Donoghue *et al* had systemic autoimmune disease such as collagen vascular diseases, thyroid disease and insulin-dependent diabetes mellitus.

Systemic immunosuppression is the most effective treatment for SINS, non-steroidals are rarely sufficient.[1,3] High dose systemic steroid therapy should be commenced promptly and tapered to a maintenance dose when healing occurs. Adjunctive therapy may include pulsed intravenous methylprednisolone, cyclophosphamide and azathioprine. Delayed use of immunosuppression has been associated with poorer visual prognosis.[1]

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